Allergy Alert

A case-based update

"I'm swollen all over!"

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Notes on Brent

Age: 13
Presentation: Presents with episodes of nonerythematous, nonpruritic swelling

- √ Episodes of swelling began during infancy, but were generally mild.
- √ Affected regions have involved lips, tongue, face and extremities.
- √ Some episodes have been precipitated by minor trauma.
- √ Most episodes of swelling have no clear precipitant.
- √ Swelling lasts one to three days.
- √ There is no residual rash, bruising or disfiguration.
- √ Episodes have become more frequent and severe over the last one to two years.
- √ Antihistamines appear to have no effect.
- √ No history of allergic rhinoconjunctivitis or asthma.
- √ No history of food allergy and no common food precipitates any of these episodes.
- √ None of the episodes have been associated with ingestion of acetylsalicylic acid or non-steroidal anti-inflammatory drugs.
- √ Patient has never taken angiotensin-converting enzyme inhibitors.
- √ No family history of atopy or swelling.
- √ Patient has no history or symptoms of lymphoproliferative disorder.
- √ Past medical history is unremarkable.
- √ He is not taking any medications and has no known drug allergies.
- √ Physical exam is normal.

What do you suspect?

Final diagnosis:
Hereditary angioedema (new mutation)

- √ Typical episodes of angioedema (nonerythematous, nonpruritic angioedema lasting one to three days).
- √ Condition presents in infancy and worsens during puberty.
- √ Minor trauma the only identifiable common precipitant, although not always present.
- √ Other common causes of angioedema are excluded.
- √ Laryngeal and gastrointestinal tract involvement possible, although not seen in this patient to date.
- √ Absence of family history does not exclude the diagnosis; structural features of this gene make it relatively prone to new mutations so that 20% of patients represent new mutations.
- √ Diagnosis is confirmed with low-serum C1 inhibitor level and low C4
- √ Treatment options include:
  1. Tranexamic acid
  2. Attenuated androgens
  3. C1 inhibitor replacement D_x

For further information, see Bowen T, Cicardi M, Farkas H, et al: Canadian 2003 international consensus algorithm for the diagnosis, therapy and management of hereditary angioedema. J Allergy Clin Immunol 2004; 114(3):629-37.

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